A Case of Non-Immun Hydrops Fetalis Due to Placental Chorioangioma

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Abstract

Choriangioma of the placenta occurs in 1% of pregnancies but it is an extremely rare condition to be large enough to threat viability of fetus or newborn due to nonimmune hydrops. A 3500 g female infant was born to a 32 year-old-woman at 36 weeks of gestation by caeserean section. Pregnanacy was complicated by polyhydramnios and preterm PROM. Apgar scores were 2 and 3 at first and 5th minutes, she had pallor, generalised edema, ascites, hepatosplenomegaly, disseminated maculopapular rash and cardiac failure. Laboratory examination revealed anemia, thrombocytopenia and leukocytosis, and increased number of erythroblasts in peripheral blood film and consumption coagulopathy. She died at 40th hours of age. Placental weight was 920 g and histopathological examination revealed a large chorioangioma. The pathophysiology of hydrops fetalis due to choriangioma and proper management during pregnancy are discussed.

Keywords: Placental choriangioma, non-immune hydrops fetalis.

Plasental korioanjioma bağlı non-immün hidrops fetalis olgusu

Plasental korioanjiom tüm gebeliklerin %1'de görülmekle birlikte fetus ve yenidoğanda fatal seyirli non-immün hidrops fetalise yol açabilmektedir. 3500 gram ağırlığında kız bebek, 32 yaşındaki polihidramniosu ve erken membran rupturü olan annenin 1. gebeliğinde 36. gestasyon haftasında sezaryen ile, 2-3 apgar ile doğdu. Soluk görünümde olan hastada jeneralize ödem, hepatosplenomegali, yaygın makülopapüler döküntüleri ve kalp yetnezliği bulguları saptandı. Laboratuar incelemesinde anemi, trombositopeni, lökositoz, periferik yaymada normoblastlarda belirgin artış ve dissemine intravasküler koagülasyon saptanan hasta takibinin 40. saatinde eksitus oldu. Plasenta ağırlığı 920 g olup, histopatolojik inceleme plasental koriyoanjiom ile uyumlu idi. Bu çalışmada koriyoanjioma bağlı hidrops patofizyolojisi ve gebelikteki tedavi yaklaşımları tartışılmıştır.

Anahtar Sözcükler: Plasentak korioanjiom, non-immün hidrops fetalis.

Background

Hydrops is generally a common end stage for a variety of diseases that share any of the three underlying mechanisms. These are the conditions that lead to congestive heart failure or conditions with obstructed lymphatic flow or decrease in plasma osmotic pressure and increase in capillary permeability.[1] In the past, most cases of hydrops were due to erythroblastosis from Rh alloimmunization, now non-immune hydrops makes up 76-87% of all cases.[2] Incidence of non-immune hydrops fetalis (NIHF) at delivery ranges from 1 in 830 to 1 in 4600.[3]

Although chorioangiomas are the most common benign tumor of placenta, hydrops fetalis due to chorioangioma is a rare condition that resulted

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from fetal cardiac failure because of hyperdynamic circulation and anemia.[4-6] We present such a case of hydrops fetalis with a review of the literature in order to emphasize the importance of follow-up in chorioangioma-detected cases.

Case

A 3500 g female infant was born to a 32-year-old unregistered gravida 1, para 0 mother at 36 weeks gestation by caesarean section. Pregnancy was complicated by polyhydramnios and premature rupture of membranes. Apgar scores were 2 and 3 at 1st and 5th minutes, respectively. In umbilical cord blood, pH was 7.18, base deficit was –10.5. On physical examination weight was 3500 g, length was 47 cm, head circumference was 35 cm. She had pallor, generalized edema and ascites. Cardiovascular system revealed tachycardia and short systolic murmur in left parasternal area. Liver was 4 cm and spleen was 3 cm palpable below the costal margin. Disseminated maculopapular rash was observed over the trunk and limbs. Laboratory examination revealed hemeglobine 11.8 g/dl, platelets 55000/mm^3 and leucocytosis 91100/mm^3 with increased number of normoblasts (60%) in peripheral blood film. Blood glucose 31 mg/dl, total bilirubin was 14.9 mg/dl with a rise in direct bilirubin of 12,02 mg/dl. AST was 785 IU/L, ALT 143 IU/L, BUN 9 mg/dl, Cr 0.4 mg/dl PT 160", PTT 82", Fibrinogen 63 mg/dl. In urinalysis 3+ proteinuria, 2+ bilirubinemia were detected. Chest X-ray demonstrated cardiomegaly with a cardiothoracic index of 0.70. She died at 40th hour of birth. In pathological evaluation placental weight was 920 g. At dissection, a mass 9x6x5 cm in diameters was seen in maternal surface of the placenta. It was purplish-red in color and well demarcated from surrounding parenchyma. Microscopically the tumor was composed of numerous blood vessels capillary in type and supported by loose, scanty fibrous stroma. No areas of necrosis, calcification or myxoid change were identified (Figure 1).

Discussion

Chorioangioma is a benign and common neoplasm of placenta that does not metastasize.[6] Fetal complications such as hydramnios, congestive

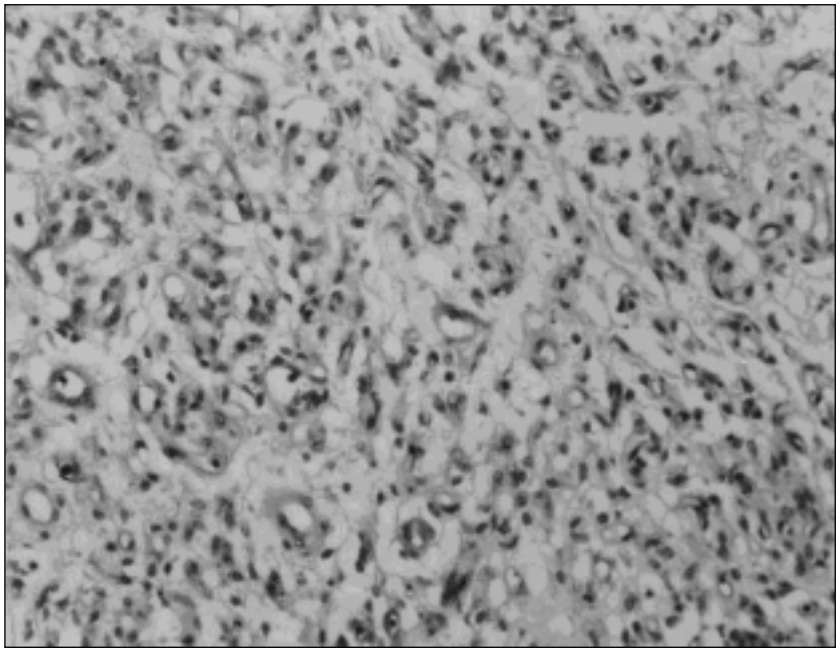

Figure 1. Numerous capillary sized vessels are separated by inconspicuous stroma (**Hematoxyline and Eosin x200**).

heart failure, anemia, and prematurity and growth retardation were rarely described.[4,7-8] Among these complications hydramnios is the most frequently reported one. When anemia develops which is severe enough to cause high output cardiac failure, fetal cardiomegaly, and hydrops can be expected. Development of anemia can be either due to hemodilution or destruction of the red blood cells in such cases.[9] Our case had congestive heart failure with cardiomegaly, hepatomegaly, edema and ascites with a hemoglobin level of 11 g/dl. Consumptive coagulopathy was also present in our case with disseminated purpuric rash and prolonged PT and PTT, low fibrinogen value and reduced number of platelets. It was suggested that trapped red cells and platelets in the vascular channels of chorioangiomas resulted in consumptive coagulopathy and microangiopathic hemolytic anemia.[10]

In most of the reported cases, it was emphasized that the size was important for the development of fetal hydrops.[7,11] However Jauniaux et al.,[12] evaluated 9 cases of chorioangiomas which in those cases the diameter of the tumors ranged between 3 and 10 cm, only one case was complicated by non immun hydrops. They concluded that the vascularization of the tumor is a pivotal determinant factor for the development of complications not the size. If the tumor is avascular no specific complications are expected. Our patient had a large placenta weighing 920 g with a large chorioangioma, which was 9x6x5 cm in size. It was also associated with increased vascularity.

With the increasing use of ultrasound, prenatal diagnosis of these tumors is becoming more common.[5] In patients with fetal and/or maternal complications, color Doppler may play a role in demonstrating the blood flow inside the mass.[4,12] As for those without complications and with little or no blood flow, it is of limited use. Management includes umbilical blood sampling and intravascular transfusion that temporarily corrects the hydrops and significantly pro- longs the pregnancy.[9] Ablation of the blood supply of placental chorioangioma via operative fetoscopy is another management alternative in patients with large chorioangioma.[13] If diagnosis cannot be made in utero or in utero treatment fails, fluid restriction with diuretics and blood transfusion is administered for the treatment of neonatal cardiac failure. Consumptive coagulopathy can be managed by fresh frozen plasma, and platelet transfusions. Life expectancy is low among non-immun hydrops fetalis cases, early in utero diagnosis of placental chorioangiomas and management will improve prognosis.

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